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Cyclic Vomiting Syndrome: The Disease in Disguise by Christine Lackey

Cyclic Vomiting Syndrome: a term I had never even heard until the fall of 2006. My then six-year-old son had preexisting GI issues since birth, but most of his problems started after he had a fundoplication done at age three for what we thought was severe reflux. We struggled for years after his surgery with weight issues, feeding issues, and motility problems, and still dealt with daily chronic vomiting. But fall 2006 brought about a new type of vomiting: cyclic vomiting.



This is our story of living with a child who has severe CVS. In the summer of 2006, my son Jake was admitted to Boston Children's Hospital for chronic pain during nighttime tube feeds. The very next day, Jake woke up vomiting. This was not typical for him, as he was usually a nighttime vomiter. He continued to vomit throughout the day. At first, everyone thought he must have picked up a GI virus, but he had no temperature or body aches. Before we knew it, a couple of days turned into a week and we were still in the hospital with Jake vomiting. We never ended up dealing with what we were there for to begin with: pain with nighttime feeds.

Over the next five months, Jake was in and out of the hospital every two weeks like clockwork with these cycles, until cyclic vomiting really started to look like his diagnosis. CVS was first mentioned by a GI attending who did a scope on Jake, but since this diagnosis is pretty much a process of elimination of other disorders that could be causing the vomiting, it took many months for Jake to be diagnosed.

Cyclic Vomiting Syndrome (CVS) is usually found in children between ages two and sixteen, although adults can receive the diagnosis as well. CVS is not very common, and thus far researchers have no definite explanation as to its cause. Symptoms include recurrent episodes of stomach pain, nausea, vomiting, and headaches. During an episode, which often appears without warning, the person can do little else but be miserable. Vomiting occurs frequently, up to 10 or more times per hour. Episodes last from a few hours to several days. Those afflicted with CVS are usually well and normal between episodes.

Because CVS is infrequently seen by most doctors, it can be very difficult to diagnose, especially since vomiting may be caused by a large number of common disorders other than CVS. There are as of yet no blood tests, x-rays, or other specific procedures used to diagnose the disorder. The diagnosis is made by careful review of the patient's history, physical examination, and imaging studies to rule out other diseases that may cause frequent vomiting. The severity of the syndrome can range from mild (not interfering with school or work) to severe and debilitating, requiring hospitalization with each cycle. The standard diagnostic criteria for CVS are listed below:

Rome III Diagnostic Criteria for CVS¹

At least 3 months, with onset at least 6 months previously of

1. Stereotypical episodes of vomiting regarding onset (acute) and duration (<1 wk).
2. Three or more discrete episodes in the prior year.
3. Absence of nausea and vomiting in between the episodes.
4. There are no metabolic, gastrointestinal, or central nervous system structural or biochemical disorders.
5. Supportive criteria: personal or family history of migraine headaches.

Although the cause of CVS is not known, most children with this condition have a family history of migraines and most experts believe the two are related. Many children find that their CVS turns into migraines during adolescence. There may also be a relationship between CVS and mitochondrial abnormalities, epilepsy, dysmotility, and several other disorders.² CVS cycles can also be triggered by a wide variety of stressors, including infections, sleep deprivation, exercise, trauma, positive or negative emotional stress, menstruation in girls, or food allergies.³

After a long and draining stay in the hospital over Thanksgiving 2006, we were left with no choice but to take Jake home still vomiting when he ran out of places for IV access. Thankfully Jake has a J-tube, so we were able to get some fluids into him when he was vomiting, but treatment with IV access has always been much more effective for him. I

went home that weekend knowing something had to give. I did some research and came across a doctor in the Midwest, Dr. David R. Fleisher, who has been working with adults and children with CVS. I left a message and surprisingly, within a day I got a phone call back. We talked for at least two hours, and I think I spent most of the time in tears because I was so desperate for help with Jake. He offered to see Jake, review his records, and try to come up with a treatment plan and a more effective ER protocol. At first I was very nervous. Insurance would not cover this trip, and we were already strapped for cash due to all the hospital admissions, none of which were even helping Jake.

Shortly after my conversation with Dr. Fleisher, Jake ended up in the hospital again for another cycle, still like clock work, to the day and time, two weeks later. Call it luck or a blessing in disguise, but the doctor that first mentioned CVS to us happened to be the GI attending on service. This doctor, Dr. Athos Bousvaros, took Jake under his wing and said, "If it is ok with you, I want to be Jake's doctor." I finally had a little bit of hope that maybe a doctor would be able to help us with this nasty CVS. This doctor stopped almost all of the 15 medications Jake had been put on prior to diagnosis and also made a very clear cut ER plan to bring every time Jake began a cycle. This smoothed the way considerably, eliminating hours of waiting and testing, and gave him immediate access to IV fluids and medication to put him to sleep.

I remember the following day feeling like a 100 pounds had been taken off my shoulders, but I still had the idea of taking Jake to go see Dr. Fleisher, a true expert in CVS. In December, our family flew out to Missouri for our six-hour appointment, knowing it was going to be nothing more than talking and reviewing his records. It was the best trip we ever made, and in the end, insurance actually covered the visit. We came home with recommended guidelines for management of CVS, as well as a recommendation that Jake get a port-a-cath, an implanted central line, due to the fact that on several occasions there were no veins available to support a regular IV for his treatment.

An important thing to know about CVS is that each cycle is characterized by four phases.⁴ These need to be noted to make sure treatment starts at the very first symptom in an attempt to "abort" the cycle from occurring. These phases include:

- Prodrome
- Episode
- Recovery
- Symptom-free interval

The **Prodromal** phase signals that an episode of nausea and vomiting is about to begin. This phase, which is often marked by abdominal pain or autonomic symptoms, can last from just a few minutes to several hours. Sometimes taking medicine early in the prodromal phase can stop an episode in progress. However, sometimes there is no warning: a person may simply wake up in the morning and begin vomiting.

The **Episode** phase consists of nausea and vomiting; inability to eat, drink, or take medicines without vomiting; paleness; drowsiness; and exhaustion. During this period, symptoms are treated or the child is sedated.

The **Recovery** phase begins when nausea and vomiting stop. Healthy color, appetite, and energy return.

The **Symptom-free interval** phase is the period between episodes when no symptoms are present.

At this point, there is no cure for CVS, but with proper treatment and experienced doctors, CVS can be managed. Preventive medications that are effective and commonly used to prevent cycles include amitriptyline (Elavil) and cyproheptadine (Periactin). For full treatment guidelines, please see this article by Dr. David R Fleisher: <http://www.articles.complexchild.com/fleisher.doc> The most important thing for anyone who suffers from or has been diagnosed with CVS is to make sure he or she is being treated...CVS will not go away on its own. The longer one suffers with CVS, the longer it can take to get the cycles under control.

The medications Jake takes for CVS are Topamax (an anti-epileptic and migraine medication), Zofran (a medicine to control nausea and vomiting), and, most importantly, Thorazine, a medication which literally knocks Jake out until the vomiting passes. Topamax has helped to space out the cycles so they occur less frequently. For Jake, Thorazine has reduced what was a week of vomiting to about 24 hours. The port-a-cath has also been a lifesaver in Jake's management for CVS, allowing easy IV access and eventually treatment at home instead of in the hospital.

It has now been 20 months since Jake began having symptoms of CVS. He has had at least 40 cycles and has been admitted into the hospital with every single cycle. Yes, 40 admissions for a seven-year-old in the last 20 months. While Jake's case is more severe than most, CVS can definitely be a debilitating and devastating disorder for any child.

For more information on Cyclic Vomiting Syndrome, visit the Cyclic Vomiting Syndrome Association [<http://www.cvsaonline.org/>].

Christine Lackey lives in Bellingham, MA with her family and is currently a stay-at-home mom of two children, Hannah, who is nine years old and in the third grade, and Jake, who is seven years old and in the first grade. Over the last couple of years, Christine has researched and talked to many doctors and other families regarding Cyclic Vomiting Syndrome. She hopes one day that there will be a cure for this disease and she continues to spread the word on how important it is for anyone who is diagnosed with CVS to make sure to get the proper treatment.

If you have any questions regarding Cyclic Vomiting Syndrome, you can email Christine at Crispyfunk@aol.com

¹ Included in Namita Pareek, David R. Fleisher, and Thomas Abell, Cyclic Vomiting Syndrome: What a Gastroenterologist Needs to Know. *The American Journal of Gastroenterology* 2007;102(12):2833.

² Pareek, 2835-6.

³ Pareek, 2834.

⁴ Fleisher, Empiric Guidelines for the Management of Cyclic Vomiting Syndrome. 2005. Available from <http://www.ch.missouri.edu/fleisher/Empiric%20%20Guidelines%20for%20Treatment%20of%20Cyclic%20Vomiting%20Syndrome%202005.pdf> A new version was released in April, 2008 and can be downloaded at <http://www.articles.complexchild.com/fleisher.doc>